Radiology Case Reports 17 (2022) 2510–2514

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Case Report

Case of an octogenarian with an asymptomatic double aortic arch undergoing CABG☆

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ARTICLE INFO

Article history:
Received 27 March 2022
Revised 14 April 2022
Accepted 15 April 2022

Keywords:
Double aortic arch
Coronary artery bypass grafting
Adult

ABSTRACT

Double aortic arch is a rare congenital anomaly of the aortic arch system where a complete vascular ring is formed around the trachea and esophagus. Case reports of elderly patients are extremely rare. We report a case of coronary bypass grafting in an elderly patient with right-dominant DAA.

Introduction

A double aortic arch (DAA) is a rare congenital anomaly where the ascending aorta divides into 2 branches, encircling the trachea and esophagus then conjoining again to form a single descending aorta. If compression of trachea and esophagus is severe, this causes significant symptoms such as dyspnea, recurrent pneumonia, stridor, dysphagia, and feeding difficulties and patients are diagnosed and treated early in life. However, when compression is minimal, DAA can remain undiagnosed until adulthood. We report an asymptomatic case with a DAA found during detailed examination of an abnormal chest shadow.

Case report

An 80-year-old male was referred to our institution for detailed examination of a small nodular shadow detected on chest radiograph at routine health examination. Chest radiograph at our institution (Fig. 1A) and subsequent computed tomography (CT) scan revealed the absence of a lung lesion, whereas DAA and abdominal aortic aneurysm (AAA) with a maximum diameter of 65 mm were incidentally detected. The preoperative evaluation for AAA revealed multivessel coronary artery disease.

Enhanced CT revealed separated right and left aortic arches (Fig. 1B) and a relatively symmetrical appearance of both common carotids and subclavian arteries (4-vessel sign) above the level of aortic arch (Fig. 2). Three-dimensional CT demonstrated the anatomy of the DAA (Fig. 3). The right sub-

☆ Competing Interests: This authors have no competing interests to report.

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https://doi.org/10.1016/j.radcr.2022.04.031

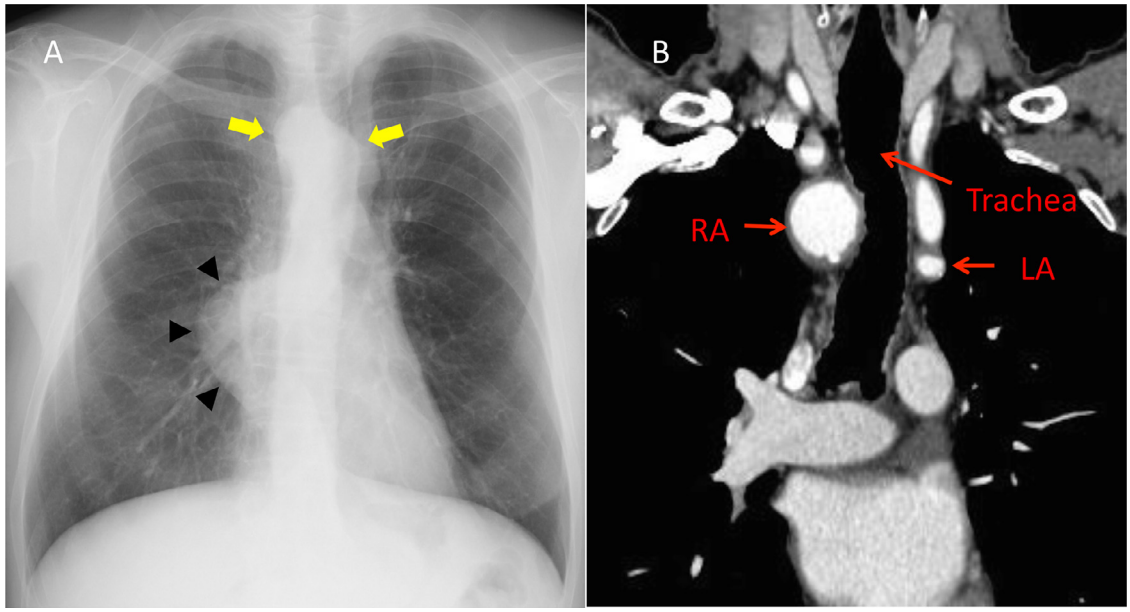

Fig. 1 – (A) Chest radiography showed the absence of a lung lesion, whereas bilateral aortic notches at the level of the aortic arch (yellow arrow), suggesting aortic arch anomaly and tortuosity of the descending aorta, projecting in to right lung (black arrow) was identified. (B) Coronal view of enhanced CT revealed separate right and left aortic arches. RA, right aortic arch; LA, left aortic arch. (Color version of figure is available online.)

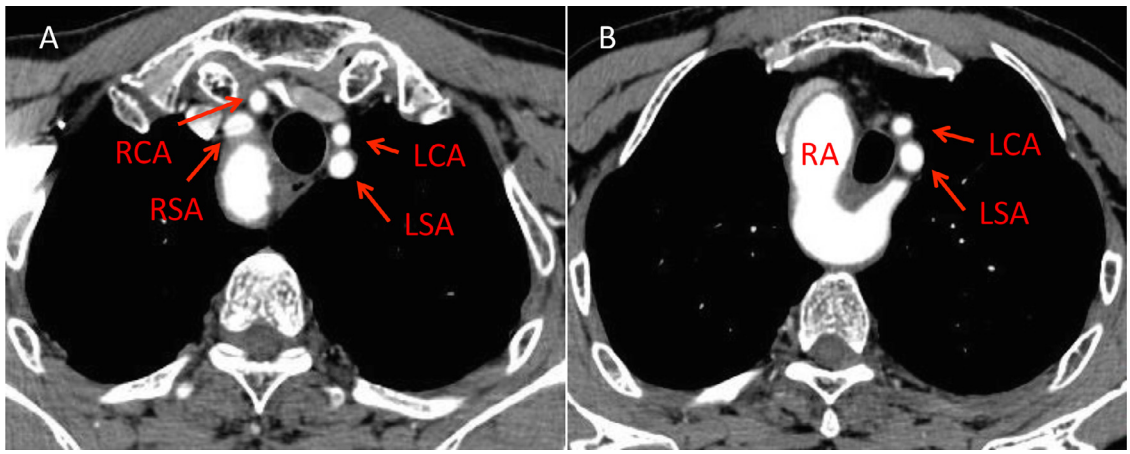

Fig. 2 – (A) In the upper mediastinum the subclavian and carotid arteries appear bilaterally symmetric. (B) At a level below A the right arch (RA) is visible to the right of the trachea. On the left side of the mediastinum the left carotid artery (LCA) and the left subclavian artery (LSA), which arise from the left arch, are visible. The trachea and esophagus are surrounded by a vascular ring made by the double aortic arch. LCA, left carotid artery; LSA, left subclavian artery; RVA, right carotid artery; RSA, right subclavian artery; RA = right arch.

clavian and right common carotid arteries originated from the right aortic arch, whereas the left common carotid and left subclavian arteries originated from the left aortic arch; the diameters of the aortic arches were 23 mm (right) and 12 mm (left). Bronchoscopy and esophagram were not performed due to the lack of significant symptoms.

We selected the staged operation of coronary artery bypass grafting (CABG) followed by endovascular aneurysm repair for AAA. Median sternotomy was performed under general anesthesia. Five vessel CABG was applied in the usual manner, by using left internal mammary artery and saphenous vein without cardiopulmonary bypass (off-pump CABG). The prox-

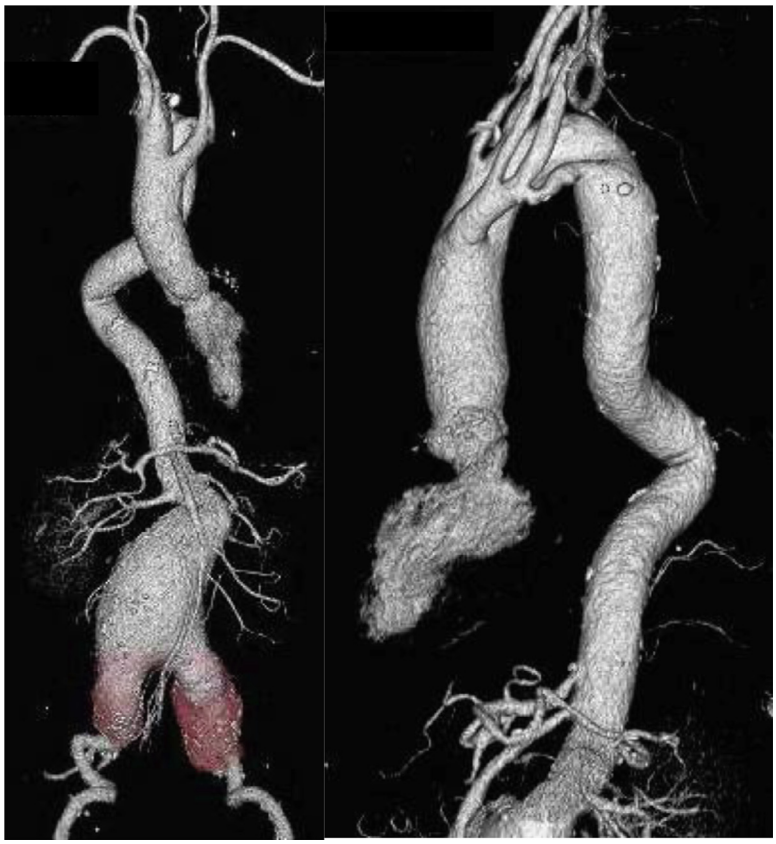

Fig. 3 – The 3D CT revealed double aortic arch separated at the distal portion of the ascending aorta and joined at the proximal portion of the descending aorta. The right subclavian artery and right common carotid artery originated separately from the right aortic arch, and left subclavian artery and left common carotid artery originated from the left aortic arch.

imal anastomoses were performed with the aid of the HEARTSTRING device (Guidant Corporation, Santa Clara, CA) to avoid side clamping of the atherosclerotic ascending aorta. Coronary bypass grafting proceeded without incident. The patient was discharged on day 11 with no neurologic sequelae.

Five weeks later, he underwent successful endovascular aneurysm repair for AAA. Postoperative CT showed good expansion of the stents without leakage of the AAA (Fig. 4) and the patient was discharged on day 6 without any complications.

Discussion

DAA is the most frequent form of vascular rings, which constitute less than 1% of the congenital heart diseases. Vascular rings were first identified by Gross in 1945 [1]. The embryonic development of vascular rings has been explained in detail by Edward's theoretical model [2]. Edwards proposed a theoretical double aortic arch system with bilateral arches and ductus arteriosi encircling the trachea and esophagus, which reflects the potential contributions of nearly all embryonic arches to components of the definitive adult arch system. In the theoretical model, both the right and the left fourth branchial arches persist leading to the so called double aortic arch, each one to be seen on either side of trachea. The anatomy of the normal left aortic arch can be described by persistence or regression of segments of the hypothetical double arch system, in contrast, double aortic arch results from lack of regression of hypothetical embryologic double arch system [3].

In the setting of DAA, bilateral common carotid and subclavian arteries arise symmetrically from the bilateral arches and the right arch is usually larger and higher than the left arch. The smaller left arch may be focally or diffusely narrowed or even atretic, with fibrous continuity or the interrupted segment. The descending aorta is typically opposite site the dominant arch, with the most common configuration being a larger right arch, left-sided descending thoracic aorta and left-sided ligamentum arteriosum.

DAA is the most common cause of a symptomatic vascular ring, as the trachea and esophagus are completely encircled and may be compressed by the 2 arches. Clinical manifestations of a double aortic arch in infants and children, usually dating from birth, include wheezing and stridor exacerbated by crying, tachypnea and cyanosis, and dysphagia. Double aortic arch is rarely associated with congenital heart disease, but when present, tetralogy of Fallot is the most common disorder, followed by transposition of the great arteries.

Diagnosis is confirmed with CT or MRI scanning which demonstrates a classic 4-vessel sign in the superior mediastinum due to separate subclavian and common carotid ar-

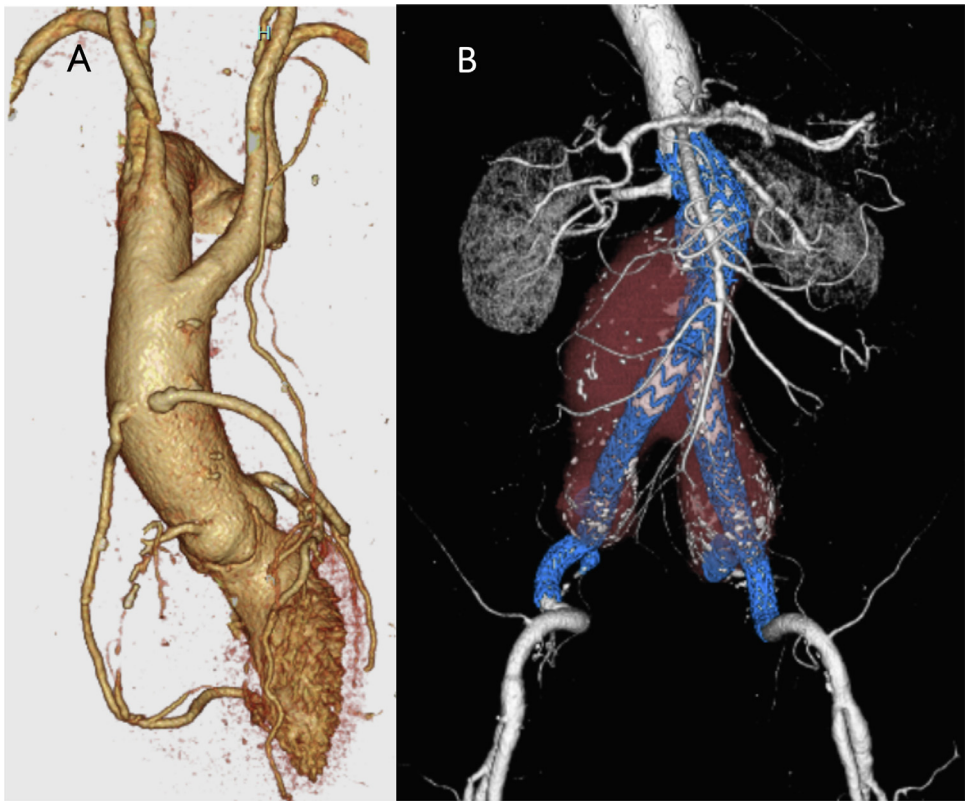

Fig. 4 – Postoperative CT scan showed (A) patent CABG grafts and (B) aneurysm exclusion without endoleak.

teries on both sides. There are three subtypes of DAA, depending on the relative sizes of the 2 arches: right-dominant (75%), left-dominant (20%), and balanced-type (5%). Echocardiography may be performed to reveal any intracardiac anomalies and may also be used for anatomical description of the vascular system. Bronchoscopy may be performed for the patients with compression of trachea to evaluate pulsatile tracheal compression. Barium esophagography may reveal esophageal compression.

Surgical repair of vascular rings is not typically indicated unless the patient is symptomatic or imaging suggests aneurysmal changes [4]. Some sporadic cases in which surgical treatments were performed in symptomatic adult patients have been reported. Saran et al [5] have described successful outcomes and symptomatic improvement in 97% of adult patients following surgery. The approach for vascular ring repair in most patients is through a left thoracotomy. Median sternotomy is reserved for patients who require tracheal reconstruction, repair of associated cardiac defects, or arch reconstruction. Right thoracotomy is used to divide a nondominant right arch in patients with DAA.

Adults with asymptomatic DAA are even rarer in literature. Data regarding asymptomatic and unrepaired vascular rings is limited. Due to the sporadic nature of this anomaly, there are not enough data on either the long-term prognosis or the standard care of untreated DAA in adulthood. A therapeutic dilemma exists when asymptomatic patients undergo surgery for another cardiac condition. Some reports document nonoperative management even in a patient undergoing coronary bypass surgery [6] with good results. Long-term sequelae of untreated DAA in patients undergoing cardiac surgery is unknown and certainly re-operation for DAA should be a significant concern.

Because our patient was asymptomatic with no compression of trachea or esophagus we did not perform concomitant surgical intervention for DAA during CABG. In case of surgical intervention for DAA, adequate mobilization of distal arch and proximal descending aorta can be safely achieved via a left thoracotomy.

In summary, we report a rare case of an octogenarian who has lived his entire life with an undiagnosed right-dominant double aortic arch that remains absolutely asymptomatic. Three-dimensional CT for DAA detection is invaluable due to its noninvasive nature and high quality images, and surgical intervention may be performed in symptomatic cases, otherwise nonoperative follow-up would be suitable.

Patient consent statement

Informed Consent Statement: Informed written consent was obtained from the patient to include the information in this publication.

REFERENCES

[1] Gross RE. Surgical relief for tracheal obstruction from a vascular ring. N Engl J Med 1945;233:586–90.
[2] Edwards J. Anomalies of derivatives of the aortic arch system. Med Clin North Am 1948;32:925–49.
[3] Hanneman K, Newman B, Chan F . Congenital variants and anomalies of the aortic arch. Radiographics 2017;37:32–51.
[4] Loomba R. Natural history of asymptomatic and unrepaired vascular rings: is watchful waiting a viable option? A new case and review of previously reported cases. Children 2016;3:44.
[5] Saran N, Dearani J, Said S. Vascular rings in adults: outcome of surgical management. Ann Thorac Surg 2019;108(4):1217–27.
[6] Ikenouchi H, Tabei F, Itoh N. Silent double aortic arch found in an elderly man. Circulation 2006;114:360–1.